Common variable immunodeficiency: etiological and treatment issues.

Journal: Int Arch Allergy Immunol

Publication Year: 2009

Authors: Sean Deane, Carlo Selmi, Stanley M Naguwa, Suzanne S Teuber, M Eric Gershwin

PubMed link: 19571563

Funding Grants: Stem Cell Research Training Grant

Public Summary:

Scientific Abstract:
One of the great advances in clinical medicine was the recognition of the pleomorphism of the immune response and the multiple afferent and efferent limbs of antigen processing and responsiveness. A significant contribution to this understanding was derived from studies of human immunodeficiency states, including both inherited and acquired syndromes. Amongst these syndromes, one of the most common, and least understood, is common variable immune deficiency (CVID). CVID is a syndrome that leads to a reduction in serum immunoglobulins and complications including recurrent infections. Management includes immunoglobulin replacement therapy; however, patients with CVID are at risk for complications of exogenous immunoglobulin administration as well as CVID-associated diseases such as autoimmune processes and malignancies. To assess the current state of knowledge in the field, we performed a literature review of a total of 753 publications covering the period of 1968 until 2008. From this list, 189 publications were selected for discussion. In this review, we demonstrate that while the molecular basis of CVID in many cases remains incompletely understood, significant strides have been made and it is now clear that there is involvement of several pathways of immune activation, with contributions from both T and B cells. Furthermore, despite the current gaps in our knowledge of the molecular pathogenesis of the syndrome, there have been dramatic advances in management that have led to improved survival and significantly reduced morbidity in affected patients.